Society Proceedings

NEW YORK NEUROLOGICAL SOCIETY.

February 2, 1904.

President, pro tem, Dr. Joseph Fraenkel, in the chair.

A Case of Huntington's Chorea (Degenerative Chorea), with Limitation of the Movements to the Face and Extremities of the Left Side.—This case was presented by Dr. J. Ramsay Hunt. The patient was a Hungarian, 33 years old, the youngest of seven children. He had been a resident of the United States for six years. No history of choreiform movements in his antecedents could be elicited. From the patient's sister it was learned that he was extremely dull, even as a child, and lacked interest in his studies. For the past three years he had twitching movements of the left upper extremity, confined chiefly to the fingers and wrist of the left hand. Similar muscle-twitching was also present in the left lower extremity, consisting of slight flexion movements in the ankle and toes. Occasionally the left quadriceps extensor contracted, elevating the patella.

The face was the seat of irregular grimaces and twitchings, and the latter were also present in the tongue, interfering at times with speech. These movements were not violent, and seemed to be increased by voluntary movements of the parts. The patient complained that the left upper extremity felt cold and heavy, and that finer movements of the hand, such as eating, were interfered with.

Mentally, the patient was dull. He was cranky and irritable at times, careless in his dress, and was unable to obtain any employment. At times, he had mild acute mental symptoms and apparent hallucinations. He had complained of frontal headaches at intervals since the onset of the trouble. Otherwise, the examination proved negative. Muscular power was unimpaired; there was no ataxia; no sensory disturbances. The pupils and all reflexes were normal. He had never had a convulsion.

Dr. Hunt said that the character of the twitchings, although slight and limited to the left side, resembled those observed in Huntington's chorea. The constancy of the symptoms, the pronounced and progressive character of the mental changes, even in the absence of heredity, pointed very strongly to a degenerative form of chorea. An initial psychosis with secondary motor manifestations would deserve consideration in the differential diagnosis.

A Case of Multiple Cerebellar Tumors.—The specimen was shown by Dr. B. Sachs. The patient was a boy, 17 years old, a native of Russia, who was admitted to Mt. Sinai Hospital on Nov. 8, 1903. He complained of vague symptoms, and was discharged after a few weeks with the diagnosis of neurasthenia.

He was readmitted to the hospital on Dec. 19th, and stated that for eight weeks previous to Nov. 8th he had suffered a great deal from occipital headache, daily vomiting, difficulty in speech and unsteady gait, associated with dizziness.

When Dr. Sachs first saw the patient, on Dec. 28th, he found, in addition to the subjective symptoms above mentioned, a very distinct scanning speech similar to that of multiple sclerosis. There were marked ataxic movements of the left upper extremity, a distinct titubating walk, and double optic neuritis. A second examination on Jan. 3, 1904, showed an increase in the ataxic tremor of the left upper extremity, which was also somewhat weaker than the right. The knee-jerks were present, but not exaggerated. There was slight ataxia of the left lower extremity; a marked plantar reflex; no Babinski sign. The scrotal reflex on the left side was increased; also the abdominal reflex on that side. There was no difference in the facial innervation, but some weakness of the left external rectus. On Jan. 8th, Dr. Gruening reported that the optic neuritis was progressing, and that there were hemorrhages in the fluid. Hearing was normal.

On Jan. 15th the patient's skull was trephined for the purpose of relieving pressure and preventing the development of total blindness. The patient died from shock a few hours after the operation.

Cerebro-Spinal Lues (Syringomyelia Symptom Complex).—This case was presented by Dr. Isador Abrahamson. The patient was a male, 35 years old; single; a peddler, and a native of Roumania. When he was 11 years old, after a fright, he became blind in the right eye and the vision of the left eye became impaired. Some improvement in sight occurred within two months.

When he was 16 years old he contracted a chancre, with the usual secondary symptoms. He was treated for this disease for one year with both external and internal remedies. He was moderately addicted to the use of alcoholic stimulants, and smoked excessively.

This case, which was reported in detail by Dr. Abrahamson, was probably one of cerebro-spinal lues, presenting the syringomyelic symptom complex, or else it was a case of syringomyelia, plus lues, or possibly of luetic origin. Such cases, the speaker thought, were rather uncommon, and were naturally of considerable clinical importance.

Dr. Sachs, in discussing this case, said he had seen some cases, which, while luetic, presented the clinical aspect of syringomyelia. He regarded these cases as examples of cerebrospinal lues, not luetic syringomyelia.

Flaccid Leutic Hemiplegia with Atrophies.—This case was also presented by Dr. Abrahamson. The patient was a male; 26 years old; married; a Russian peddler. He was an excessive smoker and used alcoholic stimulants in moderation. He had lues four years ago. Ten years ago he had an attack of parasitic alopecia, and six years ago he sustained a severe fall. This resulted in a Colles' fracture and he was said to have been unconscious for a long time.

About two months ago he was obliged to remain in bed for three weeks on account of stiffness of the neck, violent neuralgic occipital pains, and headaches, chiefly nocturnal. During the third week of this illness he began to suffer severe right-sided headaches, with vertigo.

Three weeks ago, while in a café, he felt dizzy and staggered as though drunk. He fell to the floor and found that the right side of his body was numb and paralyzed. Speech was also lost. This condition lasted about five minutes, when he apparently again felt normal. A short time afterwards he had a second attack lasting five or six minutes, and in the course of two hours he had four additional similar attacks. From the sixth attack, however, he failed to recover; speech was lost for four days and the paralysis still persisted.

At the time the case was presented there was paralysis of the lower two-thirds of the right side of the face and of the right upper and lower extremities. There was wrist-drop and foot-drop of the right side, flaccidity of the right hand and forearm; also of the leg and foot. Decided wasting was observed in the right posterior forearm musculature; also, but to a less degree, in the right leg. The electrical reactions in the posterior forearm were much diminished; myotatic irritability was diminished; sensibility was intact; heart was normal.

Dr. Abrahamson said that in view of the electrical changes, the marked wasting, flaccidity and altered myotatic irritability, all occurring within three weeks after the onset of the attack, he felt justified in calling the case a flaccid hemiplegia with atrophy.

Jacksonian Epilepsy of Luetic Origin; Improvement under Anti-Luetic Treatment.—This case also was presented by Dr. Abrahamson. The patient was a male, 39 years old, a shoemaker by occupation and a native of Hungary. He was married and the father of eight children, of whom six were living. His first wife had three miscarriages, occurring about the third or fourth month. The patient had typhoid fever in childhood. He was a heavy smoker and a moderate drinker. He positively denied venereal infection.

His present illness dated back to Dec. 15, 1903, when he began to suffer from frequent attacks which were ushered in with a feeling of numbness in the right hand, followed by a drawing in of the thumb, flexion of the fingers, and semi-pronation of the hand. The numb sensation spread to the shoulder, then down to the hip and feet. He complained of weakness of the leg and had to sit down to keep from falling. Speech was also temporarily lost. Each of these attacks, which sometimes occurred at hourly intervals, lasted about fifteen minutes, and were followed by some weakness of the right side and slight dizziness. He also complained of left-sided headaches and slight nausea and epigastric distress at the end of the attack. There was also rolling of the eyes during the attack, and rolling of the tongue upon attempting to speak. No frothing; no biting of the tongue; no diplopia were observed. Micturition and defecation were not affected. There was no loss of consciousness.

An examination showed that the right pupil was larger than the left. The pupillary reactions and ocular movements were normal. There was weakness of the entire right half of the body, including the lower two-thirds of the face. There was some exaggeration of the deep reflexes on the right side. The special senses were normal. The eyes showed a beginning papillitis.

The patient was put upon inunctions of mercury and large doses of potassium iodide. At first, the weakness on the right side increased, and a right hemianesthesia of somewhat irregular distribution developed. Subsequently, he improved rapidly, and at the time of presentation he had entirely lost his attacks, the right-sided weakness was much less, and only scattered areas of diminished sensibility were present over the right side.

Arterio-Sclerosis of Spinal Cord, with Hemorrhage.—The case was reported by Drs. Joseph Collins and Edwin G. Zabriskie. M. H., a fireman, noticed toward the end of 1902 fatigue on exertion, jerking sensations, occasional sharp pains in the lower extremities, and a sensation of heaviness, accompanied by difficulty in holding his urine. These symptoms steadily progressed, until he awoke one morning and found himself unable to stand or walk. There was also incontinence of the bowels and bladder.

On examination, it was found that he was unable to flex his thighs slowly, and there was slight atrophy of the left anterior and inner thigh muscles. The reflexes were absent; Babinski's phenomenon was present. Power in the upper extremities was fair, but he was easily fatigued. There was slight quantitative diminution of sensation in both lower extremities, and a zone of hyperesthesia as wide as the hand above the trochanters. There was slowness of speech and of mental reflexes. Also a moderate degree of general arterio-sclerosis was present. Death supervened gradually of cardiac weakness.

Autopsy: Macroscopically, there was arteriofibrosis everywhere, only slightly marked in the brain and bulb, but very marked throughout the cord. The vessels were distended with blood, and there was some extravasation into the perivascular spaces. The changes consisted in thickening of the media and propria, with loss of muscular tissue. A hemorrhage was found in the ninth dorsal segment of the cord, involving the right posterior and left anterior horns. There was sclerosis of the glia around the margin of the cord and in the lateral pyramidal tracts. Secondary degenerations were found in marginal portions of the right column of Goll, Gowers, and the direct cerebellar tracts on both sides above the lesion, and the crossed pyramidal tracts, and columns of Türck below the lesion. There was no definite degeneration of Schultze's tract or the other descending tracts of the posterior columns, excepting in the lower lumbar region, where there was a small area in the triangle of Gombault and Philippe.

Dr. Zabriskie said it was interesting, in view of the difference of opinion as to the origin of the fibers in the descending tracts of the posterior columns to note that in this case, although the right posterior horn was almost completely destroyed, the comma tract and areas of Flechsig and Philippe-Gombault were not involved; this would seem to point to the exogenous origin of these fibers.

Report of Two Cases of Tumor of the Ponto-Medullo-Cerebellar Space (Acoustic Neuroma), with Operation.—Drs. J. Fraenkel and J. Ramsay Hunt reported these cases:

Case I. Reported by Dr. Fraenkel; operation by Dr. Charles A. Elsberg. The patient was a man, 48 years old, a theatrical manager, who entered the Montefiore Home on July 1, 1903. One brother was an imbecile; otherwise his family history was negative. His previous personal history was also without bearing on the present condition. He denied excesses and venereal infection.

About five years ago the patient first noticed that his hearing on the right side was impaired, so that he had to use his left ear when at the telephone. A year later his gait became staggering and uncertain, and he could not coördinate his lower extremities at all times. The right lower extremity was the first to be affected. The patient did not notice that he swayed to one side more than to the other. Three years ago his gait frequently was like that of a drunken man, and two years ago a transient diplopia was noticed. About the same time the lower extremities became markedly spastic, his sight became impaired, his speech became stammering, and he noticed that he was tremulous when writing or performing any other intentional function with the upper extremities. During the last year, occasional insufficiency of the sphincters was noticed, and he was unable to walk without assistance. At no time was there any pain, paresthesia or girdle sensation.

An objective examination showed static and locomotor ataxia. Romberg's symptom was present. The patient fell and inclined to the left side. There was slight mental hebetude and occasional emotional explosiveness. There was motor weakness of all four extremities, particularly on the left side; a tactiform unsteadiness of both upper extremities, particularly of the right; loss of position sense in right-sided upper extremity; exaggeration of reflexes, particularly on the left side; Babinski's sign present on both sides, more marked on the left; marked nystagmus; scanning speech; considerable choked disk, with retinal hemorrhages; deafness of the left ear.

Six weeks before operation, a paralysis of all branches of the seventh nerve set in, with marked changes of the electrical reactions.

The operation for the removal of the tumor was performed by Dr.

Elsberg. The patient unfortunately died from what appeared to be an acute uremia 36 hours after the preparatory operation.

Dr. Charles A. Elsberg, who did the operation in this case, said that the opening in the skull was made in a rather unusual situation. In most of the operations for a tumor in the posterior fossa, the cerebellum was exposed by the so-called occipital craniotomy, in which the opening was made in the occipital bone somewhere between the median line and the posterior margin of the mastoid, and below the level of the lateral sinus. For tumors situated in front of the cerebellum, as was the tumor in this case, and for tumors in the anterior and lateral parts of the cerebellum there were several objections to this method of exposure:

1. It was not the shortest route from the surface of the skull.
2. The manipulations in the removal of the tumor must be done by the sense of touch alone.
3. Even with the most gentle manipulations the cerebellum was apt to be contused and its tissues lacerated.
4. In the digital examination of the anterior parts of the cerebellum from the under surface of the organ, the finger must come in frequent contact with the medulla oblongata and perhaps the pons, and the great danger from even slight injury to these structures was well known.

Dr. Elsberg said he investigated this subject for the purpose of finding, if possible, a method by which the disadvantages above mentioned would be avoided.

Actual measurements on a number of skulls showed that the shortest distance from the surface of the skull to the anterior edge of the lateral lobe of the cerebellum on the same side was measured on a line which ran along the posterior surface of the petrous portion of the temporal bone, and reached the surface of the mastoid 1 to 2 cm. behind the auditory meatus. The distance from the outer surface of the occipital bone, below the level of the lateral sinus, to the anterior edge of the cerebellum diminished steadily as the measurements were made more and more distant from the median line. These facts showed that the shortest route to the ponto-medullary space on the same side would be through an opening made in the mastoid process.

If, however, in the cadaver, an opening were made in the mastoid region and the dura incised, the lateral edge of the lateral lobe of the cerebellum fell away from the posterior surface of the petrous portion of the temporal bone, so that fully one-half of that part of the bone could be seen. With only very slight retraction, the lateral lobe could be drawn sufficiently towards the median line so that the auditory and facial nerves could be plainly seen in their course from the ponto-medullary cerebellar space to the internal auditory meatus, and even the ninth, tenth and eleventh cranial nerves could be seen in part of their course in the posterior fossa. The auditory nerve could be cut close to its origin with a fine scissors without touching the pons or medulla oblongata. Dr. Elsberg said he had performed the operation on the dead body a number of times and was convinced of its feasibility. In the living body, where the cerebellum was filled with blood, the same conditions would be found, although to a somewhat less degree. This had been demonstrated by Krause, of Berlin, in a somewhat similar operation devised by him in a case in which he cut the auditory nerve in the posterior fossa for the relief of a persistent tinnitus aurium.

As was well known, considerable of the lateral parts of the cerebellum could be cut away without causing a permanent disturbance in the functions of this part of the brain. Therefore, this was the part of the cerebellum that could be handled with the greatest safety, and at the same time could be most easily retracted. From these facts it followed that the approach to the anterior edge of the cerebellum along its outer edge was not only the easiest, but also the route by which the organ was least apt to be injured during the manipulations.

An examination into the anatomical relations of the parts in the posterior fossa would soon make it clear that when the ponto-medullary cerebellar space was approached from the mastoid region along the outer edge of the lateral lobe of the cerebellum, neither finger nor instrument need come in intimate contact with the pons or medulla oblongata until the space itself had been reached. It was probable that the correct way to remove tumors from this region was to expose the structures on the central side of the tumor, if possible, and first divide these, and then to work outwards all the time, away from the pons and medulla oblongata. Hence, with this lateral method, the danger of wounding these important structures was reduced to a minimum.

Dr. Elsberg thought it was advisable, in almost all of these cases, to operate in two stages. On Jan. 21, 1904, under chloroform anesthesia, the bone was exposed by a flap of the soft parts, including the periosteum over the mastoid and outer parts of the occipital regions. An opening was then made with the cranial drill, chisel and rongeur, measuring 5 by 8 cm., and extending from above the superior curved line of the occipital bone to within one and a half cm. of the margin of the foramen magnum, and from within one cm. of the external auditory meatus to 3 cm. from the median line. In the upper part of the wound the occipital lobe, covered by its dura, was visible, and in the lower part, the right lobe of the cerebellum, covered by its dura. The greater part of the lateral sinus of the right side was thus exposed. The wound was then closed, the flap being sutured back into place, and a voluminous dry dressing was applied.

The patient was considerably shocked at the end of the operation, but he reacted well to stimulation. His condition remained fairly good until the second day, when his kidneys ceased to act and he passed into an uremic condition and died in 48 hours.

The skin-flap was turned back about two hours after death and the dura incised. With the head turned to the side, the tumor could be plainly seen at a depth of from 3 to 4 cm. and when the finger was introduced it was found that the tumor could be peeled out with the greatest ease. It seemed probable that had the patient survived the first operation, the removal of the tumor could have been accomplished at the second operation in a short time and with very little difficulty.

After the patient's death it was learned that he had had an attack of acute nephritis, with general edema, some years before, and although the urine had been carefully examined prior to the operation and had showed no evidence of renal disease, it was just to suppose that long-standing kidney trouble was the probable cause of the acute suppression of urine after the operation.

Case II. Reported by Dr. Hunt; operation by Dr. George Woolsey. This patient was a Jewish peddler, 42 years old. Six years ago he began to suffer from an annoying tinnitus in the left ear, followed by some impairment of hearing. During the past four years he had had occasional occipital headaches, with vertigo. These symptoms gradually increased in severity and were accompanied by cloudiness of vision. For the past six months there has been an unsteadiness of gait.

When he first came under Dr. Hunt's observation, the watch tick was not perceptible with the left ear, and the tuning-fork was very faint to aerial and bone conduction on that side. There was paresis of the left seventh nerve, more apparent in its upper branch, with slight quantitative electrical changes. There was paresthesia in the sensory distribution of the left fifth nerve, with objective sensory disturbances. The pupils were unequal, the left being wider than the right. Reactions were present. There were amblyopia and choked discs, and a slow nystagmus in extreme positions. There was hemiasynergia and hemiataxia of the left side. In falling, the patient favored the left, and progression was decidedly to that side. The nuclear reflexes were active; the knee-jerks exaggerated, the left more so than the right. There was no ankle-clonus. The skin reflexes were present, but no Babinski sign.

The diagnosis of a neurofibroma of the left acoustic nerve was made, and operation was advised. From the known pathological character and relation of the surrounding structures of this variety of tumor in general, it was possible not only to localize the growth, but to state in general terms its situation in the ponto-cerebellar space outside of the brain substance, its possible size and consistency, and its enucleability. Its attachments to the surrounding structures consist only of the lepto-meninges, blood vessels and adhesions, and when these are divided it would be possible to lift the growth out of its socket in the brain with ease.

With these data at his disposal, Dr. Woolsey decided on entering the occipital bone on its postero-inferior surface, to the left of the median line and below the lateral sinus. The index finger was inserted through the trephine opening and beneath the left cerebellar hemisphere, coming in contact immediately with the nodular surface of the growth, which was broken up by gentle manipulation and removed in fragments.

There was a tendency for the cerebellar hemisphere to rise into the trephine opening, and this, with the digital manipulation, caused some laceration of its substance, with oozing.

Following the removal of the growth, the respirations of the patient became irregular and shallow, with occasional alarmingly long intervals. The pulse was rapid. The operation, which was done under ether, had required nearly two hours. At 5 P. M., three hours after the operation, the patient's temperature was 102; pulse, 112; respirations, 26. The breathing had improved in character, and the patient seemed to be reacting. Slight clonic twitchings of the left side of the face were noticeable. Death occurred at 1 A. M., apparently of respiratory and cardiac failure, just twelve hours after the completion of the operation.

Permission was obtained to remove the brain only. The outer surface of the dura was dotted here and there with little tufts and excrescences, which proved to be attached to the brain cortex, and, when torn away, they left minute depressions, with a punched-out border. Examination of the base showed that the tumor had been almost completely removed, only two small fragments remaining, together with a small blood-clot, scarcely larger than the little finger-nail. The inferior surface of the cerebellum was lacerated, but there was no external evidence of injury to the pons and medulla oblongata. The general ventricular cavity of the brain was dilated, showing that there had been a not inconsiderable internal hydrocephalus. Upon splitting the worm of the cerebellum, Dr. Hunt said he was surprised to find a firm, rounded projection from the left side of the floor of the fourth ventricle, corresponding to the site of the tumor. Microscopic sections through the brain proved this to be an extensive edema and hemorrhagic infiltration of the left peduncle of the pons, evidently arising from the region of the lacerated cerebellum, and gradually encroaching on the vital centers of the medulla. This internal pontile hemorrhage was probably the immediate cause of death.

Dr. George Woolsey said he was much interested in the class of tumors described in the cases reported by Drs. Fraenkel and Hunt, for they were so definitely localizable and could be so readily enucleated that they were very suitable for surgical treatment if they could be successfully exposed. He believed that this could be done. In the case he had operated on several months ago, which was referred to him at Bellevue Hospital by Dr. Hunt, he knew of no previous experience to serve as a guide in the operation. The tumor was approached from behind and the side by removing a large area of bone, comprising all that part of the occipital bone below the superior curved line and to the left of the median line. The opening left only a narrow bridge of bone between it and the foramen magnum and extended forward into the mastoid until the bone became very thick.

Passing the finger beneath the cerebellum, the tumor was felt and removed piecemeal. This necessitated introducing the finger a number of times, and there was some laceration of the lateral lobe of the cerebellum which did not seem of any moment. The tumor was larger than he expected to find it. In another case he would try to pass a forceps along the finger, seize the tumor, enucleate it with the finger and remove it entire.

The plan of operating in two stages, recommended by Horsley and employed by Dr. Elsberg he thought was most excellent where the exposure was prolonged and much blood was lost. He thought Dr. Elsberg's method of approach had much to commend it, although it was not so much nearer the site of the tumor than the opening he had made as might be inferred. It had the objection that the bone was very thick, so that too much time and perhaps blood was lost in getting through it. In operations on brain tumors it was important not to take too much time in the exposure alone. This objection was obviated by operating in two stages. Dr. Woolsey said the entire operation in his case took no longer, if as long, as the exposure in Dr. Elsberg's case.

After the operation, his patient seemed in good condition and appeared to suffer but little from shock, but symptoms of the latter appeared several hours later, and he failed to rally. On the whole, in another similar case, he would be inclined to operate in two stages and by the lateral approach suggested by Dr. Elsberg.

Dr. George E. Brewer said that he had been interested in the subject of cerebral tumors for many years. He had twice explored the posterior fossa, in the first instance finding a subcortical cerebellar growth of tuberculous origin, which was successfully removed, the patient making an excellent operative recovery. He died some months later, however, from general tuberculosis.

In the second case the patient succumbed after an extensive bone removal, undertaken simply to relieve pressure. A number of anatomical investigations carried on in the dissecting room of the College of Physicians and Surgeons led the speaker to advocate the plan of opening the skull by removing, practically, all of the bone below the superior curved line to within half an inch of the foramen magnum below, and from the median line to a point half-an-inch behind the mastoid. By opening the dura, the finger could be easily passed behind the lateral lobe and reach the situation of these tumors at a depth of less than an inch and one-half from the surface of the wound. Of course, adequate exposure to remove such a growth would necessitate the destruction of a portion of the lateral lobe: this, however, could be done without serious trouble to the patient, as had been demonstrated in his first case.

It was Dr. Brewer's opinion that speed of operation was perhaps the most important factor necessary to success, as these patients almost invariably died if the operation was at all protracted. He was in favor of doing the operation, when practicable, in two stages. The first operation should consist of the removal of the bone only, which to a certain extent relieved the pressure, and at a subsequent period the dura should be incised and the tumor removed. He thought the primary operation should be done in from twenty to thirty minutes.

Dr. Hunt, in closing, referred to a previous communication, published in conjunction with Dr. Fraenkel, in which five cases of tumor of the ponto-cerebellar space were recorded. In that paper, especial stress was laid on the certainty of localization and the enucleability of this group of tumors, and their surgical importance.

In both of the cases under discussion, in which operation was attempted, the diagnosis of an acoustic neuroma was made, and the operative procedure was modified to meet the known anatomical peculiarities of this type of tumor. In case I the patient succumbed 36 hours after performing the first stage of the operation, apparently from shock, although a renal complication could not be excluded. In case II death was due to laceration of the cerebellum, with resulting effusion of blood into the tissues of the medulla oblongata and pons. Both of the operations were prolonged and tedious, and considerable trauma was inflicted in removing the thick occipital bone, and enlarging the trephine opening. It seemed of paramount importance, in exposing tumors in this locality, that the operation be done expeditiously and with great gentleness, as any concussion must be especially effective in this region. The division of the operative procedure into two stages was also to be recommended.